TWO CASES
OF
CONGENITAL MACROSTOMA

ACCOMPANIED BY

MALFORMATION OF THE AURICLES AND BY THE
PRESENCE OF AURICULAR APPENDAGES.

BY

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ALTHOUGH I have been unable to find any description of exactly parallel cases to those which are here detailed, they will be found to bear out the observations of previous writers on cases of a somewhat similar character, and to add to our knowledge of the appearances which may result from vestiges of rudimentary portions of the foetal branchial apparatus.

CASE 1.—Maud Bailey, æt. 13 months, was brought to me when two months old. Her parents had been married seven years, and had had three healthy children, all of whom are alive, and free from deformity of any sort. The strictest inquiry fails to elicit an account of abnormality in any member of the families on either side. The child was very small when born, and for some weeks

did not thrive, but has made fair progress up to the present time, though it is still far from being a well-grown baby. No history of any value can be obtained as to any accident or personal injury occurring to the mother during the earlier period of her pregnancy.

The deformity was obvious, and the following peculiarities were observed when the child was first brought to the Children's Hospital.

The formation of the mouth upon the right side was natural, but upon the left its aperture extended into the cheek in a direction backwards and slightly downwards towards the angle of the jaw. The fissure by which it was prolonged was fully three quarters of an inch long, and involved the whole thickness of the cheek. It was covered with mucous membrane similar to that of the lips, and continuous with the skin of the cheek. The spot where the lips should have been united was indicated by a small papilla on either side of them, but owing probably to the absence of the entire orbicularis muscle this spot was further from the middle line on the deformed than on the natural side. Within the mouth the gum of the lower jaw was more prominent than upon the opposite side, whilst from beneath the tongue there protruded a mass of vascular mucous membrane, having much the appearance of a ranula, and which was more pronounced upon the left than upon the right side.

In the left external ear it was observed that the space between the tragus and antitragus was carried downwards towards the angle of the oral fissure and lying upon the surface of the cheek, and placed in this line were two small oval growths, one attached by a single, and one by a double pedicle covered with skin, resembling that of the tragus, in which there are small, fine hairs. No cartilage could be felt. The external auditory meatus was situated in front of the normal position, and running slightly backwards, became narrowed at its extremity, which was filled with the usual secretion of cerumen. Downwards and forwards from this ran another passage,

which terminated in a *cul de sac*, and was about half an inch in length. The lower maxilla appeared to be considerably smaller than it should have been, but seemed to be bony throughout. The movements of the lower jaw were not symmetrical.

On the right side of the face was a small growth similar to those on the left cheek, and situated in front of the tragus. The condition of the meatus resembled in a less marked degree that described as existing on the opposite side.

During the last twelve months the child has increased in size, and the condition mentioned remains the same, although the size of the fissure appears still more exaggerated.

So far as can be ascertained hearing is equally good upon both sides, and examination of the meatus (although unsatisfactory from its narrowness) does not reveal any further abnormality.

CASE 2.—Elizabeth Monro, æt. 5. The parents are Scotch, and are healthy people. They have one older child, a boy, aged 9, who is weakly, but perfectly well formed. Two children were born between him and the subject of the present notice; one, a boy, died at birth, the other, a girl, died at twelve months old of whooping-cough. No deformity was noticed in either of them, nor can it be ascertained that any has existed in members of the families of either parent.

This child was very delicate at birth, and is said to have had the valve of the heart unclosed, and for some time to have been in a very critical state. She has since suffered from abscesses, which have made her delicate.

The left side of the mouth is natural, but the margin of the right extends into the cheek for about three quarters of an inch, and the edges of this fissure are covered with mucous membrane similar to that of the lips, and continuous with the skin of the cheek. Two small papillary elevations exist at the spot on each lip

where they should be united, and these are made more or less evident by muscular action, as though marking the insertion of fibres of the orbicularis muscle. There is a faint line, as of a slight cicatrix, which runs from the external angle of the fissure over the surface of the cheek towards the external meatus. No trace of this is seen on the mucous membrane, which differs only from that of the opposite side by being rather more anterior at the part which unites the upper and lower jaws. There does not appear to be any difference between the two sides of the inferior maxillæ, and although the whole bone is somewhat smaller than usual, the number of teeth upon both sides is equal. In both ears the external auditory meatus at its outer part is broad, and the space between the tragus and antitragus is wide and prolonged downwards. No marked abnormality is evident in the rest of the channel or the ossicula so far as can be observed.

Two elongated oval appendages, covered by hairy skin, are placed in a line running from the external meatus towards the angle of the fissure on the right cheek, to which they are attached by a pedunculated base, and two somewhat larger but similar growths are seen lying in the same relation on the left. The child hears equally well with both ears, although not quite perfectly with either, and she talks naturally.

Comparing the two cases, it is to be noticed that although elongation of the mouth is on different sides it is accompanied by very similar phenomena. Both cases occur in girls, which confirms the observation of Heusinger that deformities in connection with the branchial apparatus are more frequent in females. In neither case is there the history of any occurrence during pregnancy which would account for any arrest or even deviation of development; but contrary to what has been observed in cases of branchial fistula, there is no history of any hereditary tendency to aberration. The unclosed portion of the mouth extends in each case to the margin of the bucci-

nator, and although in both children this abnormal condition exists upon one side only, it is in each case accompanied by a deviation from the normal condition of both ears. While, also, the aberrant formation of the mouth points rather to deficiency, that of the ears and their appendages show, on the other hand, a tendency towards excess of growth. This alone sets at rest any suggestion as to the possibility of the deformity being due to any intra-uterine pressure or ulceration.

Very few congenital deficiencies of the body occur unilaterally, the majority being either symmetrical or in the median line. This is more especially so in the face, and the 'Teratological Catalogue' in the College of Surgeons' Museum does not contain a single specimen of asymmetrical deformity, whilst but few are recorded in the literature of this subject.

Looking to this fact, I think that the origin of the deformity of the mouth must be sought for, not in an error of formation of the mouth itself, which is developed by depression of the outer surface of the embryo above the first branchial arch, but to a failure of union between that upper part of the first visceral arch from which is developed the superior maxilla and that lower part of the same arch from which is developed Meckel's cartilage, upon which the lower jaw is formed. And this is more likely to be the case from the ill-developed condition—very marked in the one case—of the inferior maxilla, as well as by the presence and position of the "auricular appendages." Similar appendages, however, are not mentioned as existing in the only similar cases of elongation of the mouth. One of these is described by Sir W. Fergusson, where the fissure seems to have closely resembled that described in these two cases; and the second is figured in Von Ammon's 'Atlas,' and the plate is copied by Vrolik, the fissure being represented as extending upwards on the right side nearly to the angle of the eye. These are the only two cases in which this deformity has been noted.

Of the "auricular appendages," the origin suggested by Sir James Paget in his article on "Branchial Fistulæ in the External Ears,"¹ is no doubt the correct one, namely, that they are aberrant remnants of the opercular skin-fold of the first post-oral branchial cleft from which the natural auricle is developed. This opinion was coincided in by Professor Parker, to whom I showed the second of these cases. They are doubtless identical with similar growths which have been noticed by many authors as associated with branchial fistulæ, although there is no recorded case in which they have been observed in similar positions, and are said by Heusinger to be much more frequent in all domestic animals than in man. The same author states that the skin metamorphoses, as remains of the closure of the cleavings, are not unfrequently met with in the form of cicatrix-like striæ, small pore-like openings, folds of skin, and small warty-looking pieces of skin before the ears and on the throat.

Dr. Urbantschitsch, who met with twelve cases of fistulæ in 2000 patients, did not, however, once come across one of these growths. In the only case of branchial fistula which I have observed, and which occurred in a boy, and was situated in front of the right sterno-mastoid, there were no such excrescences. But I have seen one case of a girl in which they existed in front of both ears as small pendulous growths in front of the tragus; and in a case of a boy in whom the right external meatus was closed and the whole auricle small and deformed, there existed a small appendage in front of the tragus of the left ear, which was otherwise naturally developed.

The cases mentioned by Dr. Allen Thomson bear a much closer analogy to the two just described, since in one of them a deformity of the ear in a young woman "was accompanied by considerable imperfection in the shape of the lower part of the face; the lower jaw was remarkably short, its position oblique, and its angle very

¹ I have to acknowledge my obligations for references both to the article, and to Sir James Paget himself, who saw these cases with me.

obtuse. The malar bone was placed far back on the cheek, and the zygomatic arch much shorter than usual. In this instance two sisters showed deficient development of the ear.

Authors Quoted.

Sir James Paget, "Branchial Fistulæ in the External Ears" (Med. Chir. Trans., vol. lxi, pp. 41—50); Sir William Fergusson (Progress of Anatomy and Surgery, p. 56); Dr. Allen Thomson (Edin. Monthly Journal of Med. Science, vol. vii, April, 1874); Dr. Heusinger (Deutsche Zeitschrift für thierische Medicin und vergleichende Pathologie, B. ii); Dr. Urbantschitsch (Edinburgh Med. Journal, Feb., 1878, p. 691), Von Ammon's Atlas, pl. IV.

Appendix.

Since reading the above paper, I have been informed of the death of the child whose condition was first described. This occurred from an attack of bronchitis, and I was unfortunately not able to obtain a post-mortem examination.

I have operated on the second child for the closure of that portion of the mouth which was abnormally prolonged and for the removal of the auricular appendages. The latter were dissected off and the former was closed by a plastic operation with a most satisfactory result, it being at the present time difficult to detect that any deformity had existed, and it is interesting to note that the action of the orbicularis has so far improved that but little difference is discernible on the two sides of the mouth when that muscle is in action.

I have since seen one other case, which, as regards the deformity, resembled those described above in every particular, the prolongation of the mouth being on the left side, but remarkable on account of its occurring in a boy, the second child of healthy parents, whose first child is a

well-formed girl, and who can speak accurately as to the absence of any similar deformity in any member of either family for three generations. The photograph of another boy presenting an almost identical deformity to that occurring in the second case described above has been kindly sent to me by my friend, Mr. F. A. Southam, of Manchester. In this case also there was no family history of abnormality.

DESCRIPTION OF PLATE II.

Congenital Macrostoma with Malformation of Auricles and with Auricular Appendages (JOHN H. MORGAN M.A., F.R.C.S.).

CASE 2.—Eliza M., aged 5 years.

Fig. 1.—Congenital macrostoma and auricular appendages of right ear.

Fig. 2.—Auricular appendages of left ear.